Collagen is the most abundant protein in the human body.

It provides the structural strength in most human tissue, including the heart and blood vessels, eyes and skin, cartilage and bone.

What happens when this basic building block is flawed?

When muscles, ligaments, tendons and even large organs are built with structurally defective collagen there is systemic weakness and instability evident throughout the body.

There is Ehlers-Danlos Syndrome.

Excessively mobile joints, chronic pain and skin softness characterize Ehlers-Danlos syndrome (EDS). At least six types of EDS have been identified; clinical manifestations vary according to type and may also include poor wound healing with atrophic scars, easy bruising, chronic pain and generalized connective tissue fragility. Each type is thought to involve a unique defect in connective tissue, although not all of the genes responsible for causing EDS have been found. Within each family the type of EDS runs true, but individual family members may vary in clinical severity and manifestations.

As a group of genetic disorders of connective tissue, the estimated prevalence of EDS is 1 in 5,000. It is known to affect men and women of all racial and ethnic backgrounds.

Chronic pain is a well-established and cardinal manifestation of EDS and it is common for pain to be out of proportion to physical and radiological findings. The etiology of EDS pain is not clearly understood, but some of the likely causes include muscle spasm (tender points are sometimes present) and degenerative arthritis; neuropathic pain is also common.

A cardinal feature of EDS is pain. At first there are just small pains; but acute pains may accumulate, then become continual and chronic. There may be a major dislocation or injury to start the pain cycle. Without adequate treatment, persistent pain can change the nervous system in a process that is difficult to reverse; by lowering the threshold for pain signals, chronic pain becomes harder to treat.

Probably the most important element of a successful therapeutic intervention for EDS is bringing that pain under control. When a person with EDS reports pain, even if out of proportion to physical findings, they should be listened to and believed.

Percent of EDS Population

<table>
<thead>
<tr>
<th>Statistics</th>
<th>Percent</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chronic pain +6 months</td>
<td>90%</td>
</tr>
<tr>
<td>Worsened over lifetime</td>
<td>84%</td>
</tr>
<tr>
<td>Chronic pain preadolescence</td>
<td>89%</td>
</tr>
<tr>
<td>Taken pain meds</td>
<td>88%</td>
</tr>
<tr>
<td>Taken narcotics</td>
<td>53%</td>
</tr>
</tbody>
</table>

Statistics from Chronic Pain as a Manifestation of the Ehlers-Danlos Syndrome (Sachetti et al, 1997); full text available at ednf.org.

We all pay when pain is untreated

Inadequately-treated pain affects everyone:

- Productivity is decreased at work and in everyday tasks at home
- Hospital stays are lengthened and ER/clinic visits increase
- Health care costs increase
- Quality of life worsens for those in pain and for those around them:
  - Physical cost, appetite loss and weakness
  - Psychological cost: depression and anxiety
  - Emotional cost: suffering and anger
  - Social cost: lost relationships
  - Loss of independence, self-worth and financial stability can be an ultimate price

ednf.org
INCREPENT PAIN EVENTS (less than four days per week)

- ANALGESICS
  - May be ineffective as single agents or require excessive doses
  - Short-acting opioids
  - Muscle relaxants

- PHYSICAL THERAPY
  - Reconditioning
  - Exercise (gradual toning for stability, not strength)

- PSYCHOLOGICAL THERAPY
  - Relaxation
  - Stress management (incl. Mindfulness-Based Stress Reduction)
  - Group therapy
  - Cognitive restructuring (incl. hypnosis)

- SLEEP THERAPY
  - Sleep disturbance is common in EDS (contributes to poor pain recovery; incl. apnea)
  - Tricyclic antidepressants (TCAs)

- COMPLEMENTARY/ALTERNATIVE
  - Meditation
  - Acupuncture
  - Massage
  - Yoga

- MAINTAIN GENERAL HEALTH
  - Avoid weight extremes & smoking

CHRONIC PAIN (constant & disturbing pain)

- ANALGESICS FOR CHRONIC PAIN
  - Use combinations of medicines on a schedule to keep pain under control

- PREVENT RECURRING INJURY
  - Hypermobile joints and fragile connective tissues endure chronic injury
  - Bracing may be helpful (maintain toning exercise)
  - Orthopedic surgery should be avoided
  - Almost never provides long-term improvement

- OCCUPATIONAL THERAPY
  - Delay disability and encourage activity as long as possible
  - Body mechanics/ergonomics
  - Intervention in workplace and work simplification
  - Pacing skills

- NEUROPATHIC PAIN
  - Common in EDS, also called neuralgia (different from usual nociceptive pain); described as numbness, tingling, burning, electrical, hot/cold
  - Incl. dysfunctions of peripheral nerves (numbness, weakness), nerve injury, axonal loss

- NON-PAIN PATHOPHYSIOLOGY
  - Side effects from treatments, incl. overuse or ineffectiveness of analgesics or short-acting opioids

- LONG ACTING OPIOIDS
  - Methadone, morphine, oxycodone, fentanyl, oral/rectal/pump
  - Tolerance for pain-killing builds up slowly, but not to side effects incl. GI dysmotility & reduced consciousness

- SURGICAL PAIN INTERVENTION
  - Injection (trigger point, nerve root)
  - Implantable nerve stimulators
  - Medication pumps (subcutaneous cavity)

TOLERANCE, DEPENDENCE & ADDICTION

Tolerance is not addiction.

- The capacity to endure continued subjection to something, esp. a drug; diminution in the body’s response to a drug after continued use.

Physical dependence is not addiction.

- A state of adaptation that includes tolerance and is manifested by a drug class specific withdrawal syndrome that can be produced by abrupt cessation... Physical dependence on and tolerance to prescribed drugs do not constitute sufficient evidence of psychopharmacological substance use disorder or addiction. They are normal responses that often occur with the persistent use of certain medications.

The choice may be simple: dependence on drugs or dependence on pain.

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  - The fact or condition of being addicted to a particular substance, thing, or activity, when a person has lost control over and continues use even when such use is doing them or others harm.

The chance of addiction is very low when these medications are taken as directed by a doctor and used for pain.

URGENT INFORMATION on Vascular EDS

- Arterial rupture is the most common cause of sudden death.
  - Painful arterial injury is being a part of a particular substance, thing, or activity, when a person has lost control over and continues use even when such use is doing them or others harm.

Partners Against Pain*

Keeping you informed about new developments in pain management.

In 1993, Purdue created Partners Against Pain® (PAP) to forge an informational alliance among physicians, nurses, pharmacists, hospice personnel, pain experts, patients, and caregivers in the fight to ease cancer and non-cancer pain. Purdue sponsors the award-winning website — www.partnersagainstpain.com — an invaluable resource that provides pain information, assessment tools, and support 24 hours a day.

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